CASE REPORTS

Recurrent Parotitis with Sialoangiectasis

IRVING L. WHITE, *M.D.*, Long Beach

CHRONIC RECURRENT PAROTITIS is reported with increasing frequency in children and young adults. Often it is generalized throughout the gland and it may be unilateral or bilateral. Usually sialoadenitis (and rarely fistula formation) is present. Recurrent parotitis in most cases is associated with sialoangiectatic changes. Duct or alveolar ectasia may result from acquired blockage[5] due to calculus or inflammatory changes; but the ectasia may also be due to a congenital developmental structural deformity antedating the manifestation of sialoadenitis. Since infection may complicate congenital sialoangiectasis, it is often impossible to differentiate between primary and secondary disease.

Sialoangiography is an excellent method of demonstrating sialoangiectasis, and correlation with histologic findings is sometimes possible. As excision of a biopsy specimen of the parotid gland may be a hazardous procedure, it is justifiable only if the information anticipated is important enough to outweigh the danger of damage to the facial nerve.

Submitted May 31, 1962.

The author had opportunity to consider the relationship of ectasia to recurrent infection in the course of treating siblings who had chronic recurrent parotitis and sialoangiectasis. The patients were two sisters, in each of whom symptoms of recurrent parotitis with sialoangiectasis developed at the age of 17. One of them had bilateral disease although the symptoms were unilateral.

REPORT OF CASES

CASE 1. A 21-year-old white married woman was first observed June 14, 1961, with a history of recurrent swelling in the right preauricular area for over three years. The swelling would persist for one to three days and could be relieved by exerting pressure over the parotid gland. At the onset, attacks occurred at intervals of two to four months, but they had progressively increased in frequency until at the time the patient was seen by the author they

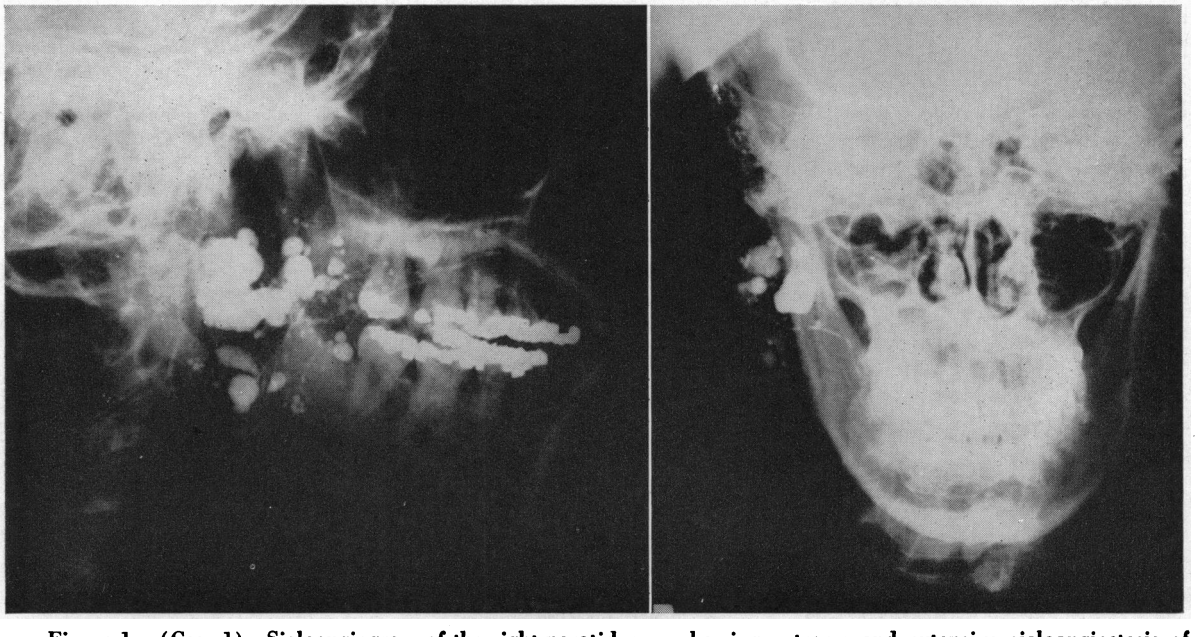

Figure 1.—(Case 1)—Sialoangiogram of the right parotid area showing extreme and extensive sialoangiectasis of the right parotid gland with saccular dilatation of acini as large as 1.5 cm. in diameter. The duct does not appear distended, nor is there any evidence of ductile obstruction. These changes are characteristic of chronic parotitis with pronounced saccular dilatations of acini elements, compatible with congenital sialoangiectasis.

were occurring every two to three weeks. An x-ray of the parotid area a year previously had not shown any calculus or radiopaque material in the parotid area.

There was no history of any previous illness. Tonsilloadenoidectomy at the age of three was the only surgical treatment she had undergone. Specifically there was no history of any swelling or soreness of the left parotid gland. The patient was four months pregnant when first seen.

Extensive questioning about family history of symptoms referable to the parotid area elicited a report of swelling of the face in the patient's sister (Case 2, hereafter reported).

Upon physical examination the only positive significant finding was tender diffuse right preauricular swelling over an area some three or four centimeters in diameter with involvement of the right parotid gland. Light pressure on the enlarged gland expressed a sero-purulent discharge from the right Stensen's duct. Probing of the duct facilitated milking of the gland. Sialoangiography showed extreme and extensive sialoangiectasis of the right parotid (Figure 1). During the next four weeks, the patient had two additional attacks of purulent sialoadenitis.

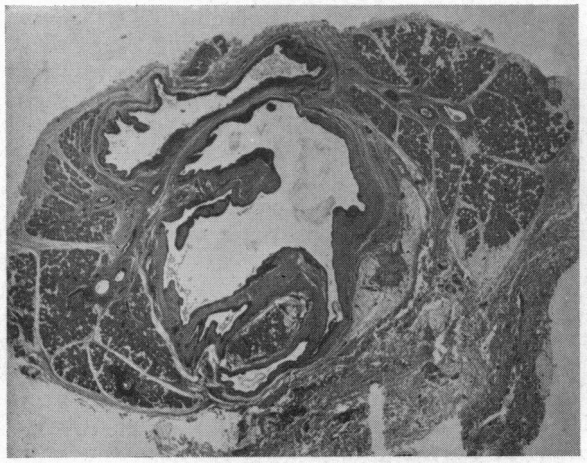

Figure 2.—(Case 1)—Portion of salivary gland showing ductal ectasia (×5).

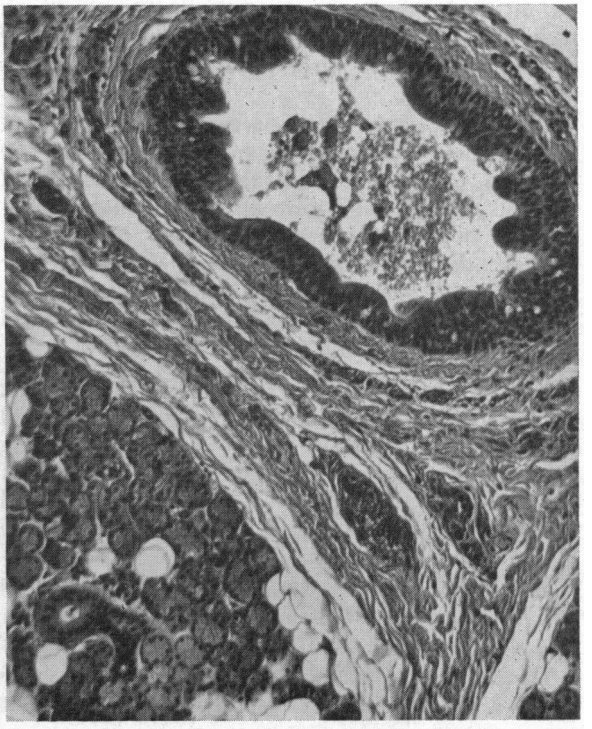

Figure 3.—(Case 1)—Salivary gland with ductal epithelium intact but showing some "piling up" of the cells (×200).

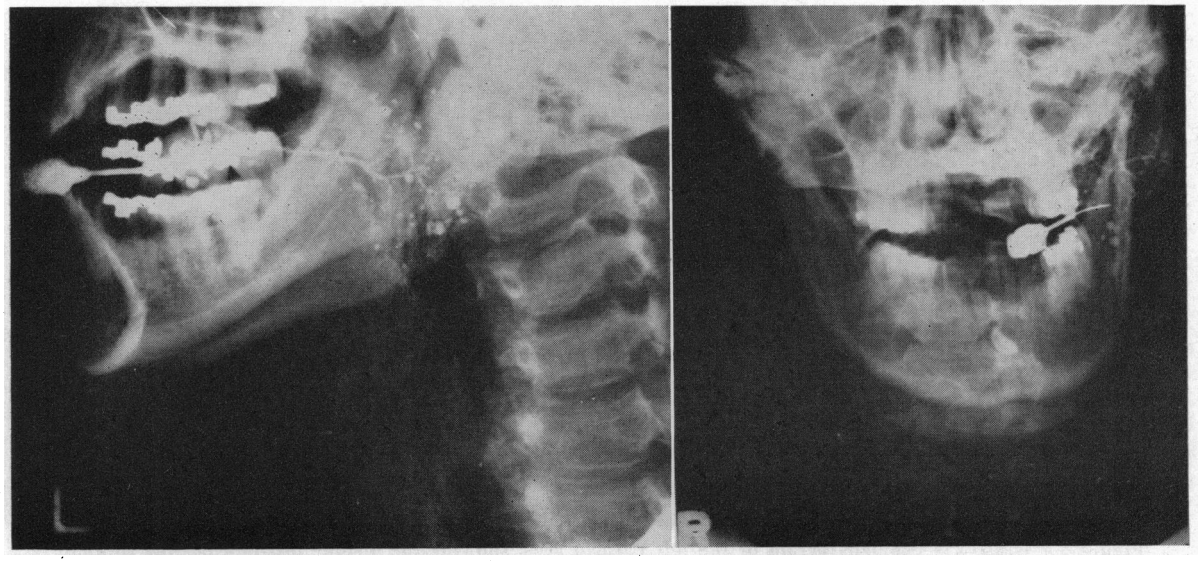

Figure 4.—(Case 1)—Sialoangiogram of left parotid area, showing sialoangiectatic changes with saccular dilatations adjacent to the non-dilated ducts throughout the parenchyma of the gland.

Because of the obvious irreversibility of the pathologic process and the increasing frequency of attacks, it was decided that operation should be done before the third trimester of pregnancy. Total parotidectomy with preservation of the facial nerve was carried out. The glandular tissue was nodular with a multitude of cysts on the surface as well as throughout the parenchyma.

Upon microscopic examination pronounced dilatation of the duct system, extending to the finest radicals, was noted. The findings were typical of sialoangiectasis and chronic sialoadenitis. (See Figures 2 and 3.)

The postoperative course was completely without complication. Following normal termination of the patient's pregnancy, sialoangiographic studies of the left parotid gland were made. Although no history of left parotid swelling could be elicited, studies demonstrated extensive sialoangiectatic changes with saccular dilatations of ducts and in the parenchyma adjacent to ducts in the left parotid gland (Figure 4).

CASE 2. The patient, a 20-year-old white woman, a sister of the patient of Case 1, was first observed June 14, 1961, with a history of cystic swelling behind and below the left ear for over two and a half years. During that time incision and drainage had been carried out more than six times, with release of bloody purulent discharge.

The patient was asked about but could not recall any swelling of the preauricular areas of either side. She had had tonsilloadenoidectomy at age 2.

Upon physical examination an inflamed cystic lesion 2 cm. in diameter behind and below the left ear was observed.

A sialoangiogram (Figure 5) showed the parotid duct posteriorly entered a dilated structure below and behind the left ear. There were dilated structures along the superior and medial aspects of the cystic structure as well as small but discrete sacculations along the duct ramifications, with no dilatation of the duct. The findings were consistent with sialoangiectasis and fistula formation.

Left total parotidectomy was carried out. The postauricular cyst and fistula were excised en masse and the facial nerve was preserved. The pathologic diagnosis was sialoangiectasis with abscess formation and chronic sialoadenitis.

The patient recovered without complication. No evidence of sialoangiectasis was observed later in a sialoangiogram of the right parotid gland.

COMMENT

In reports dealing with pathogenesis of sialoangiectasis of the parotid gland, the importance of recurrent infection has been stressed. Pathologic studies done by Jones[3] convinced him that dilatation of the finer ducts led to stagnation of secretion, which favored development of infection. Fifteen cases of sialoangiectasis, five of which were bilateral, were reviewed by Bisgard and co-workers.[2] In that series there was no obstruction to any part of Stensen's duct, ruling out dilatation of the duct being secondary to ductile obstruction. Perzik[6] demonstrated that changes in chronic parotitis associated with fistulae, sialoadenitis and sialoangiectasis are similar, since all are due to duct and glandular alterations, the main variation being one of degree. He observed that duct blockage, a complicating factor in practically all inflammatory processes, may be partial or complete, intermittent or continuous.

Planka,[7] in an exhaustive review of the literature, showed sialoadenitis to be a definite clinical and pathologic entity of the newborn, as well as of nurslings and very young children.

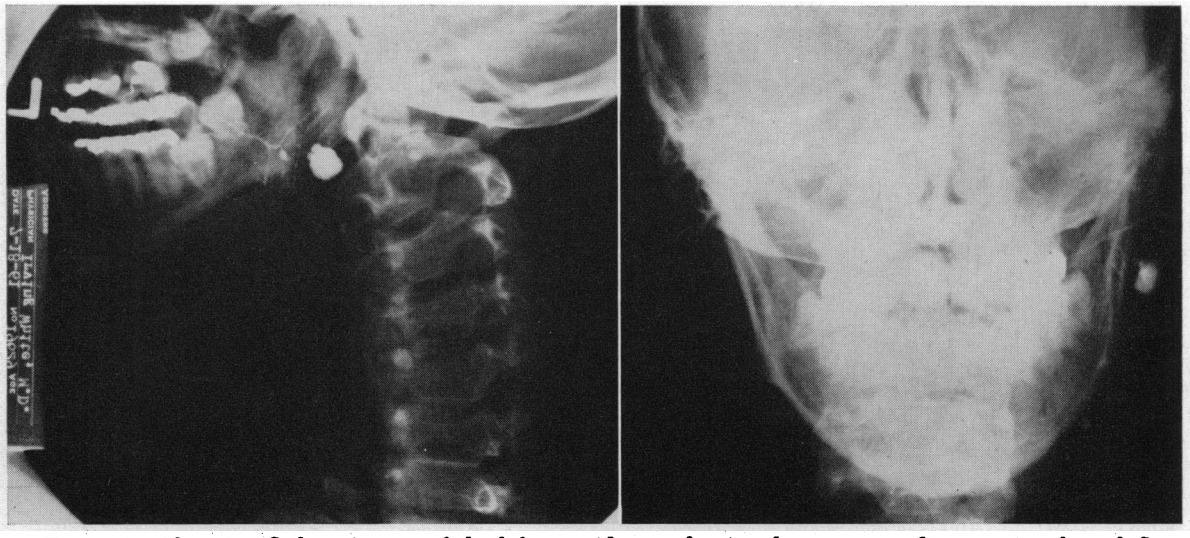

Figure 5.—(Case 2)—Sialoangiogram of the left parotid area, showing the opaque medium passing through Stensen's duct posteriorly to enter the dilated fistulous cyst. There are small but discrete dilated sacculations along the duct and adjacent to the cystic structure, but no dilatations of the duct.

Krepler[4] reported the case of a 2-year-old boy who at the time of a primary attack of pyogenic parotitis was shown by sialoangiography to have ectasia of the duct on the diseased side and small alveolar dilatation on the clinically normal side. Kepler also presented summaries of cases of a 5-year-old boy and a 16-month-old girl, each of whom had unilateral acute sialoadenitis for the first time. In those cases sialoangiography within four weeks after the attacks demonstrated sialoangiectatic changes in both sides. Krepler expressed belief that the fact that sialoangiectasis is demonstrable early in the course of recurrent sialoadenitis, often with the first attack, coupled with the young age of these patients, supports the postulation that congenital abnormalities are predisposing factors. It was on the basis of sialoangiographic pictures that he proposed the term *congenital sialoangiectasis of the parotid.*

Jones[3] reviewed 20 cases of recurrent parotitis in childhood, including cases in which the mother of two siblings with recurrent parotitis had had recurrent parotitis as a child.

Becker and co-workers[1] observed that in most cases of chronic parotitis sialoangiograms show saccular dilatation of ducts throughout the whole parotid gland, and that ectasia is always demontrable in both parotids even though the clinical disease may be confined to one.

If the pathologic processes are severe enough to warrant, total parotidectomy with preservation of the facial nerve offers an excellent form of management of sialoangiectasis and sialoadenitis.

SUMMARY

A clinical review of two sisters who had recurrent parotitis with sialoangiectasis is presented with a correlation of sialoangiographic studies and histopathologic findings. One of the patients had fistula formation behind and below the ear as a complication of the disease. In one of the two cases sialoangiographic studies showed typical pathologic changes on both sides although clinically the disease was unilateral.

The etiologic interrelationship of recurrent parotitis and sialoangiectasis is discussed. The evidence supports the presumption that congenital sialoangiectasis may be a basic underlying factor in many cases of recurrent parotitis.

911 East San Antonio Drive, Long Beach 7.

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The Mongolism-Leukemia Syndrome

Report of a Case

ROBERT A. BLOSSOM, *M.D.*, Ventura

A NINE-MONTH-OLD NEGRO BOY was admitted to hospital because of severe anemia. The obstetrical record showed that he had been born after 38 weeks of gestation. At birth he weighed 5 pounds 1¾ ounces, was 17¾ inches long and was cyanotic and responded sluggishly. He was considered to be borderline premature. By the ninth postpartum day the infant had intense jaundice and the liver and spleen were enlarged. Total serum bilirubin reached a maximum of 15.5 mg. per 100 cc. (direct, 7.8 mg. per 100 cc.) by the twentieth postpartum day. During this same period the hemoglobin dropped from 11.5 gm. to 8.0 gm. per 100 cc. A battery of laboratory tests were done in an attempt to determine the cause of the jaundice. Among those with negative result were: Coombs test (direct and indirect), serial blood cultures, liver function tests including cholesterol determination, cephalin flocculation and thymol turbidity. Preparations for demonstration of sickle cells (sodium bisulfite method) were strongly positive on four occasions. A discharge diagnosis of sickle cell anemia with crisis was made. In the ensuing months, after discharge, the child was seen for infant diarrhea and for upper respiratory infection. No additional icteric episodes were observed, although sickle cell preparations were consistently positive.

When admitted again at nine months of age, the child was noted to be decidedly anemic and a mongoloid diathesis was apparent: Slight slanting of the eyes, flattened palpebral fissures with prominent epicanthal folds, bradycephalic head and large deposits of fat in the cheeks. There was a moderate disproportion in the size of the upper and lower jaw. The mouth was small, the tongue large and protruding. Hypoplasia of the phalangeal bone caused shortening of the fourth finger. The palm was square and across it was a single crease running parallel to the line of insertion of the fingers. The entire body musculature was hypotonic and all joints were easily hyper-extendable.

At this time the sclerae were clinically icteric and the liver and spleen were palpable. Hemoglobin con-

Submitted August 8, 1962.

From the Department of Pathology, University of Southern California School of Medicine, Los Angeles 33.